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## CONGENITAL CARDIOLOGY SOLUTIONS (PEDIATRIC CARDIOLOGY AND ADULT CONGENITAL HEART DISEASE)

# THE ROLE OF IMAGING THE DISTAL AORTA IN ADULTS WITH MARFAN SYNDROME

ACC Poster Contributions
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Session Title: Aortic Issues in Marfan Syndrome and Adult Congenital Heart Disease
Abstract Category: Adult Congenital Heart Disease
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**Background:** Dissection of the ascending aorta remains the most common cause of death in adults with Marfan syndrome (MFS). As such, patients undergo serial echocardiograms to assess for aortic root dilation. Imaging of the distal aorta (dAo) is not universal. We sought to determine the frequency of dAo disease in a group of adults with MFS and to look at clinical correlates of dAo disease.

**Methods & Results:** Retrospective chart review of all adults (age > 18 years) followed between 1999-2009 in a Marfan Cardiology Center was performed. Of 176 adults with MFS, 121 had undergone > 1 MRA/CTA of the dAo. These patients formed the study cohort. Of the 121, 36 (30%) had aneurysms of the dAo or its branches. Within this group of 36, 14 had prior replacement of the ascending aorta, and 22 (61%) had not. Within the nonsurgical group of 22, 23 aneurysms were found: 7 carotid, 2 cerebral, 2 aortic arch, 2 subclavian artery, 6 in the descending thoracic aorta, and 4 in the abdominal aorta or its branches. Among the 14 surgical patients with dAo disease, 7 (50%) had dAo aneurysms contiguous with the previous repair site and 50% had aneurysms separate from this site. Of the 36 patients with dAo aneurysms, 24 (66%) experienced dissection of same. Age at dAo dissection was 33 + 10 years. DAo aneurysms were found on routine screening in 18 (50%) and at the time of symptomatic dissection in 50%. Patients with dAo aneurysms did not differ from those without, in gender (55% vs 59% female, p=0.07), presence of lens dislocation (22% vs 30%, p=0.04), aortic root diameter (47+10 mm vs 44 + 9 mm, p=0.2) or family history of MFS ( 77% vs 73%, p=0.6). Patients with dAo disease were older (41+13 vs 34+12, p=0.006), more apt to smoke (25% vs 4%, p=0.002), have hyperlipidemia (33% vs 9%, p=0.001), have a family history of aortic dissection (47% vs 23%, p=0.02), not have received treatment with an ACE inhibitor (2% vs 52%, p<0.0001) and have undergone aortic root replacement (69% vs 45%, p=0.02). Mortality rate was higher in those with distal disease (17 vs 1%, p<0.0001).

**Conclusions:** DAo aneurysms are common in adults with Marfan syndrome even in the absence of prior aortic surgery. Periodic imaging of the distal aorta should be part of routine surveillance for all adults with MFS.